REFLEX SYMPATHETIC DYSTROPHY: CRITERIA FOR DIAGNOSIS AND TREATMENT * † ‡

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The goal of therapy is to interrupt the progress of disordered physiological and pathological processes and to assist the natural reparative mechanism in restoring integrity of structure and function. To accomplish this goal, certain conditions must be fulfilled: The nature of the disordered process must be clear, the efficacy of the therapeutic method must be established, and the method of treatment must affect directly and with certainty the causative or perpetuating mechanisms of disease. These ideal circumstances are encountered but rarely in clinical practice, yet they present a challenge to all concerned with treatment.

The problem of reflex sympathetic dystrophy has been such a challenge previously because of vagueness of definition, uncertainty of diagnosis, and confusion arising from failure to recognize that many previously described entities are actually variations of one basic disease process.

This syndrome may actually be subdivided into three grades of severity, and we believe that accurate diagnosis and classification is the cornerstone of effective therapy. Our experience in the treatment of 160 cases has provided the foundation for our criteria of classification, has given prognostic significance to these criteria and has, in general, guided the choice of therapeutic management.

DEFINITION

Reflex sympathetic dystrophy may be defined as a disordered response by an extremity to trauma. Trauma ordinarily initiates and elicits certain typical responses. Immediately there is pain, altered circulatory dynamics and restriction of function. As healing progresses, pain subsides and disappears, circulatory homeostasis reappears, function, consistent with the extent of injury, is restored, and wound healing progresses toward anatomical integrity. The speed of these processes is dependent upon the nature and extent of injury, the presence or absence of infection and general metabolic or nutritional diseases, and the adequacy of treatment.

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In contrast to this orderly process of restitution and repair, the
disordered response of reflex sympathetic dystrophy is characterized
by persistent or unduly severe pain, unrelenting aberrations of periph-
eral vascular circulation, delayed or incomplete restoration of func-
tion, and a variety of trophic changes involving the entire extremity.
This chaotic response is, in our experience, completely unrelated to the
severity of trauma and is not constantly associated with injury to any
specific tissue. There are no organic constitutional states which pre-
dispose to its occurrence and there appears to be no means of fore-
telling, at the time of trauma, in which direction the reparative response
will proceed.

This syndrome, which has in part been described previously under
various names (causalgia, minor causalgia, Sudek’s atrophy, post
traumatic dystrophy, and so forth) actually is one entity which is
characterized by four constant manifestations: pain, vasomotor dis-
turbances, delayed return of function and trophic changes (1). All of
these must be present, in greater or lesser degree, in order to satisfy
the rigid criteria for the diagnosis of reflex sympathetic dystrophy.
Certain variations may, however, be observed. The pain may be ex-
erciating and constant, or it may be dull, aching, throbbing and pres-
ent only upon motion. As a rule, it is not limited to a single dermatome
nor to a specific area of nerve distribution, but may possibly be limited
to a distinct area of sympathetic nerve distribution as yet not clearly
delineated anatomically. Vasomotor changes may be mild or severe.
Vasoconstriction may predominate and the skin of the extremity
will be cold, damp and glistening, or excessive vasodilatation may be
present and the skin will be warm, dry and scaly. Trophic changes are
constant but are variable in degree and extent. Early, edema is com-
monly present; but as the disease pursues its uninterrupted course,
atrophy of the skin, subcutaneous tissue, muscle and tendon, and
osteoporosis of bone, develop. Pain may inhibit function early in the
disease but, as trophic changes develop, motion is mechanically re-
stricted and this dysfunction may be the principal permanent result
of the disease.

Grades of Severity

We have been able to define and detect three distinct gradations of
severity in reflex sympathetic dystrophy based on mode of onset, in-
tensity and relative predominance of one group of symptoms over the
others. These distinctions, so important in defining therapy, are based
on careful appraisal of symptoms and physical signs and are in no way
related to severity of trauma nor to injury of specific tissue.

Grade 1. The dominating clinical features in this group are severe,
burning, knife-like and lancinating pain, unrelieved by rest and ag-
gravated by the slightest emotional or physical disturbance. The
onset as a rule occurs within a few hours after injury but occasionally
may be delayed for days or weeks. Vasomotor disturbances are of
either type, vasoconstriction or vasodilatation; these occur early and are obvious and severe. Motion is voluntarily and absolutely restricted and atrophy of soft tissues soon appears and becomes pronounced.

Grade 2. This is a milder form of reflex sympathetic dystrophy, slower in development and longer in duration since the symptoms do not present an insistent demand for therapy. Pain is usually dull, throbbing, aching and diffuse; it is aggravated by motion and relieved by immobilization and rest. Vasomotor disturbances are mild but are constantly present and result in either peripheral vasocostriction or vasodilatation. Edema may appear early, but atrophy soon develops, persists and may result in irreversible damage to soft tissue and bone. Osteoporosis is common. As a result of these trophic changes, and because of the increased pain on motion, the patient voluntarily restricts activity early in the course and later finds that motion has become restricted organically because of mechanical factors.

Grade 3. This, the mildest form, is the most common in our experience (table 4). These patients represent the border zone between the normal response of an extremity to trauma and the more severe disorders previously described. We have observed this variation most commonly after surgical procedures upon the extremities. It is our belief, however, that because of the mildness of the symptoms and paucity of physical signs, many patients in this category never received the benefit of diagnosis and appropriate therapy. The onset is early and insidious and may be first manifested by complaints of pain in excess of that which experience commonly associates with the trauma in question. In fact, pain is usually the dominant symptom. This pain persists and prevents or retards the institution of passive or active exercises. Occasionally, when the extremity has been immobilized in plaster of Paris bandages, it becomes necessary to remove the casings; yet, upon removal, no evidence of excessive pressure or constriction is observed. Vasomotor changes are present, both objectively and subjectively. The patient experiences temperature changes limited to the extremity, which may be cold and damp or dry and warmer than the uninvolved side. Edema surrounding this site of operation or trauma appears early and is persistent. Trophic changes are mild or moderate, slow in appearing, but are constantly present; they are eventually troublesome since function will be restricted as a result.

Initiating and Sustaining Factors: the Sympathetic Nervous System

The exact etiology of reflex sympathetic dystrophy is unknown and one can only speculate upon the fact that identical trauma may, in one individual, initiate an orderly response, and in another, may initiate a completely disorderly and chaotic response. It has been established that the metabolic response to trauma is initiated, at least in part, by
the sympathetic nervous system, and it is known that with the same
degree of trauma, individual responses may vary (2). By analogy,
one may assume that in an extremity, trauma may set up afferent
sympathetic as well as somatic impulses of different intensities. One
may also postulate, and the experimental work of Bingham (3)
is confirmatory to a degree, that there are definite variations in the
number, distribution and type of sympathetic nerve fibers in the ex-
tremities. At any rate, with afferent impulses of greater intensity
entering the spinal cord, a spread of excitation occurs and synapses
are formed between sympathetic and somatic afferent fibers and central
efferent fibers of all types. This may result in the increased pain and
the vasomotor disturbances of reflex sympathetic dystrophy. In ini-
tiating these mechanisms, we are certain that the sympathetic system
plays an integral part, as a result of stimulation of afferent pain and
proprioceptive fibers which are probably present in greater number
than in the average individual.

The perpetuating and sustaining factors in this syndrome are at
least three in number: [1] the variations in individual cerebral integra-
tion and response to pain; [2] the relationship between the hypo-
thalamus and the cerebral sympathetic centers; and [3] the activity
of the internuncial pool. It seems certain that preformed cerebral pat-
terns of response vary from individual to individual. Identical stimuli
will usually produce responses which resemble these preformed pat-
terns and are only slightly related to severity of the stimulus. Fur-
thermore, hypothalamic reactivity may also vary, and since the con-
nection between hypothalamus and cerebral sympathetic centers are
numerous, this may account for variations in the perpetuation of the
response. White (4) has called attention to this hypothalamic factor
in patients with severe forms of reflex sympathetic dystrophy. Finally,
variations in the activity of the so-called internuncial pool, described by
DeNo and elaborated by Livingston (5), are significant in accounting
for variations in the extent of the spread of excitation and in the number
and types of reflex mechanisms which perpetuate the syndrome and act
as a vicious cycle.

The exact pathogenesis of reflex sympathetic dystrophy would seem
to depend, therefore, on variations of sympathetic nerve distribution,
defects in central integration and response, variations in activity of the
hypothalamic sympathetic pathways and the internuncial pool.

THE PHYSIOLOGICAL BASIS OF TREATMENT

The common denominator in all three grades of reflex sympathetic
dystrophy is involvement of the sympathetic nervous system. The exact
nature of this involvement is obscure, yet the ubiquity of sympathetic
nerve fibers is well recognized; these may be afferent pain or proprie-
ceptive fibers, the so-called nocifenser nerves of Head, or they may be
efferent fibers mediating vasoconstriction and dilatation (6). Further-
more, injury to large or small mixed somatic nerves may set up extra spinal reflexes or cross stimulation effects on sympathetic fibers, as postulated by Doupe, Cullen and Chance (7).

Whatever mechanism is involved, it appears certain that pain and vasomotor disturbances are the two factors present initially which produce the subsequent dysfunction and trophic changes. As has been pointed out, voluntary restriction of motion occurs because of pain which is increased by movement. Alterations in circulatory dynamics also appear to cause the trophic changes which soon appear and are related in severity to the extent of circulatory disturbances. On this basis, interruption of sympathetic activity appears to be physiologically sound. Clinical experience has verified this hypothesis, since interruption of this activity gives almost immediate succor from pain and restores circulation to normal.

In Grade 1 sympathetic activity is almost frenetic. It is constant, severe and unrelenting and all associated phenomena are similarly severe. Physiologically, it appears that the organism has reached the peak of sympathetic activity from the cerebral centers and the hypothalamic-sympathetic pathways down through the medullary, spinal and extraspinal ares. It would appear that nothing short of complete and permanent sympathetic denervation can halt this process permanently since the initiating factors have set up a self-perpetuating vicious cycle. Therefore, interruption by ganglion block, while perhaps providing temporary amelioration for the duration of the block, probably will not eliminate the vicious cycle which has been set up.

Patients with reflex sympathetic dystrophy of lesser severity (Grade 2) present a difficult problem in the choice of therapy: Sympathetic activity is severe but not unrelenting. Perpetuating mechanisms are not so firmly established, and perhaps the spread of excitation in spinal and cerebral centers is less extensive. It has therefore seemed logical to believe that a period of prolonged continuous, sympathetic block lasting from fourteen to twenty-eight days might be effective in overcoming the perpetuating factors of the disease by allowing local healing of the traumatized area to occur (8). During the period of blockage, pain is relieved, circulation becomes normal and, as a result, motion can be instituted and trophic sequelae minimized.

In cases of moderate severity and relatively short duration, this method may suffice. However, it is probable that a certain number of patients will develop recurrence of symptoms after termination of the block and will require surgical sympathectomy.

In the mildest form (Grade 3), diagnosis should be established early in the course. Careful observation during the immediate postoperative or post traumatic period will usually elicit the four classical signs and symptoms of reflex sympathetic dystrophy in their incipiency and while trophic changes are vascular rather than structural. In such cases prompt institution of continuous paravertebral procaine
ganglion block will interrupt the process and allow relatively uneventful healing. In this mild form, only the initiating factors are present; that is, sympathetic overactivity or cross stimulation and fixed cerebral and hypothalamic pathways. The vicious cycle has not yet developed. The effectiveness of the continuous block method of sympathetic interruption is directly related to the interval between onset and therapy. It is necessary to stress the fact that patients in this group may be classed as neurotics or malingerers unless careful examination of the traumatized extremity is performed frequently, since the signs, while always present, may be minimal.

TREATMENT AND RESULTS

If the foregoing physiological postulates are valid, clinical experience should supply the empirical verification. In our experience with

<table>
<thead>
<tr>
<th>Condition Which Was Followed by Evidence of Reflex Sympathetic Dystrophy</th>
<th>No. Cases</th>
<th>Results of Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Posttraumatic, upper extremity</td>
<td>15</td>
<td>Excellent* 1 Good** 12 Poor*** 2</td>
</tr>
<tr>
<td>Posttraumatic, lower extremity</td>
<td>1</td>
<td>Excellent* 0 Good** 1 Poor*** 0</td>
</tr>
<tr>
<td>Postherpetic, lower extremity</td>
<td>2</td>
<td>Excellent* 0 Good** 2 Poor*** 0</td>
</tr>
<tr>
<td>Totals</td>
<td>18</td>
<td>Excellent* 1 Good** 15 Poor*** 2</td>
</tr>
</tbody>
</table>

* Complete and persistent relief of symptoms and return of function.
** Adequate and sustained relief of symptoms for duration of block.
*** No apparent relief.

the treatment of 160 patients suffering from reflex sympathetic dystrophy this has been established. In 1952 we reported briefly on the results of treatment of 84 patients (8). Of this group only one patient, suffering from the severest form of the disease (Grade 1), was relieved completely or permanently by continuous procaine paravertebral block. At the other extreme, 48 of 49 patients with the mildest form (Grade 3) either recovered completely or were markedly improved by continuous paravertebral block (table 5). Subsequent experience with 76 additional patients has further corroborated the physiological postulates expressed (table 4).

The results of treatment in Grade 1 patients is summarized in table 1. Although continuous procaine paravertebral block was used in all cases in this group it served merely as temporary palliation, and in so doing, confirmed the diagnosis and classification. Of 15 patients, only one was completely relieved by the block, whereas while 12 had some
TABLE 2
RESULTS OF CONTINUOUS SYMPATHETIC NERVE BLOCK ON PATIENTS
WITH REFLEX SYMPATHETIC DYSTROPHY, GRADE 2

<table>
<thead>
<tr>
<th>Condition Which Was Followed by Evidence of Reflex Sympathetic Dystrophy</th>
<th>No. Cases</th>
<th>Results of Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Excellent*</td>
</tr>
<tr>
<td>Shoulder-hand syndrome</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>Minor injury</td>
<td>27</td>
<td>17</td>
</tr>
<tr>
<td>Fracture, old</td>
<td>12</td>
<td>7</td>
</tr>
<tr>
<td>Postoperative status, old</td>
<td>7</td>
<td>2</td>
</tr>
<tr>
<td>Phantom limb and stump pain</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td><strong>Totals</strong></td>
<td><strong>57</strong></td>
<td><strong>27</strong></td>
</tr>
</tbody>
</table>

* Complete and persistent relief of symptoms and return of function.
** Incomplete symptomatic relief although some improvement of function, or complete relief for duration of block.
*** No symptomatic relief or functional improvement.

measure of relief for the period of sympathetic blockade, the symptoms recurred promptly upon termination. Two patients in the group received no benefit from the continuous paravertebral block. This confirms our belief that all patients with the severest form of reflex sympathetic dystrophy should be subjected to surgical sympathetic denervation without delay, since delay increases the probability of residual dysfunction and perpetuates pain patterns.

TABLE 3
RESULTS OF CONTINUOUS SYMPATHETIC NERVE BLOCK ON PATIENTS
WITH REFLEX SYMPATHETIC DYSTROPHY, GRADE 3

<table>
<thead>
<tr>
<th>Operative Procedures</th>
<th>No. Cases</th>
<th>Results of Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Excellent*</td>
</tr>
<tr>
<td>Bone fusions</td>
<td>27</td>
<td>21</td>
</tr>
<tr>
<td>Fractures (simple)</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Closed reduction</td>
<td>15</td>
<td>14</td>
</tr>
<tr>
<td>Open reduction</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>Fractures, compound</td>
<td>8</td>
<td>5</td>
</tr>
<tr>
<td>Crush injuries</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Saucerization for osteomyelitis</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Mid-thigh amputation</td>
<td>8</td>
<td>6</td>
</tr>
<tr>
<td>Tendon repair</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>Nerve repair</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Blood vessel repair</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>Pedicle skin grafts</td>
<td>85</td>
<td>63</td>
</tr>
</tbody>
</table>

* Complete analgesia; no edema; return to normal circulation; early motion; accelerated healing.
** No analgesics necessary; minimal edema; return to normal circulation and earlier functional return.
*** No apparent relief.
Reflex Sympathetic Dystrophy

While surgical sympathectomy was considered advisable for all patients in Grade 1, and was performed on 10 of the 18 patients (D. F. C.), it is much less often necessary in those patients with disease of lesser severity (Grade 2). Again, continuous procaine paravertebral block was instituted in all, but, in this group, it was with hope of permanent benefit. Of 57 patients treated, continuous block gave excellent results in 27 cases. These patients were completely relieved of pain, circula-

<table>
<thead>
<tr>
<th>Reflex Sympathetic Dystrophy</th>
<th>No. Cases</th>
<th>Excellent</th>
<th>Good</th>
<th>Poor</th>
<th>Success, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade 1</td>
<td>18</td>
<td>1</td>
<td>15</td>
<td>2</td>
<td>47</td>
</tr>
<tr>
<td>Grade 2</td>
<td>57</td>
<td>27</td>
<td>26</td>
<td>4</td>
<td>70</td>
</tr>
<tr>
<td>Grade 3</td>
<td>85</td>
<td>63</td>
<td>19</td>
<td>3</td>
<td>85</td>
</tr>
<tr>
<td>Totals</td>
<td>160</td>
<td>91</td>
<td>60</td>
<td>9</td>
<td>75</td>
</tr>
</tbody>
</table>

* Rating: Excellent = 100%; Good = 50%; Poor = 0%.

tion and function returned to normal and trophic sequelae were minimal. Upon termination of the block after periods of from fourteen to twenty-eight days, improvement was maintained and no further therapy was necessary. In 26 cases, results of procaine blockade were good; that is, pain was markedly relieved and function was improved. However, in some of these, relapses occurred upon termination of the block.

<table>
<thead>
<tr>
<th>Reflex Sympathetic Dystrophy</th>
<th>No. Cases</th>
<th>Excellent</th>
<th>Good</th>
<th>Poor</th>
<th>Success, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1</td>
<td>9</td>
<td>0</td>
<td>8</td>
<td>1</td>
<td>44</td>
</tr>
<tr>
<td>Type 2</td>
<td>26</td>
<td>12</td>
<td>11</td>
<td>3</td>
<td>67</td>
</tr>
<tr>
<td>Type 3</td>
<td>49</td>
<td>34</td>
<td>14</td>
<td>1</td>
<td>84</td>
</tr>
<tr>
<td>Totals</td>
<td>84</td>
<td>46</td>
<td>33</td>
<td>5</td>
<td>74</td>
</tr>
</tbody>
</table>

* Rating: Excellent = 100%; Good = 50%; Poor = 0%.

In 4 cases no relief was obtained. Of the entire group, 11 (19 per cent) were subjected to surgical sympathetic ganglionectomy (D.F.C.). We believe that all patients in this category should have an adequate trial of continuous procaine block, since a high percentage will be completely and permanently cured. It seems evident that the results of this type of management depend upon the duration of the disease and the extent of structural damage (table 2).
The results of treatment of Grade 3 patients by continuous para-
vertebral block were almost uniformly satisfactory (table 3). Early
diagnosis and prompt institution of therapy resulted in complete and
permanent relief in 63 cases (74 per cent) and in a rapid return of
functional integrity with only minimal sequelae in 19 (22 per cent).
In only 3 cases was this type of therapy without value and in these the
condition pursued a slow and indolent course. No patient in this
group was considered for surgical sympathetic denervation. These
excellent results can be attributed to early diagnosis, the result of
alert clinical observation. With early therapy directed at prolonged
interruption of sympathetic activity by continuous procaine block, a
vicious cycle is prevented and the deleterious effects of prolonged pain
and vasomotor disturbances are minimized or completely prevented.

SUMMARY AND CONCLUSIONS

The application of rigid diagnostic criteria based on comprehension
of the physiological mechanisms involved is necessary for the success-
ful therapy of reflex sympathetic dystrophy. The ultimate goal is
restoration of complete functional and anatomical integrity of the ex-
tremity at the earliest possible time and by the simplest therapeutic
procedures. To achieve this, diagnosis must be implemented by ac-
curate classification which, on the basis of the experience presented,
affords a guide for the selection of the therapeutic method: continuous
procaine paravertebral block or surgical sympathetic ganglionectomy.

All patients suffering from reflex sympathetic dystrophy should
be benefited by proper therapy, and many will be cured. Selection of
the method of treatment best suited to the individual case at the
earliest moment will increase the number of cures and reduce the
number of permanent sequelae resulting from long continued vascular
disturbances and from disuse due to pain. The classification of reflex
sympathetic dystrophy herein presented offers such a prognostic and
therapeutic guide and enables one, at the time of diagnosis, to proceed
with therapy with a definite concept of the probability of success
and failure.

Our experience indicates that surgical denervation is necessary for
all patients in Grade 1, whereas continuous paravertebral block will
be efficacious for all patients in Grade 3. These patients in the inter-
mediate zone present the most difficult problem. In these a trial of
therapy by continuous paravertebral block is mandatory as the first
therapeutic measure, and this will probably be definitively successful
in 80 per cent of the cases. For the remaining 20 per cent of patients
who did not respond satisfactorily, surgical sympathectomy will be
necessary. Procrastination beyond a month is unwarranted since, if
no relief has been obtained by blocks in that period, there is small
likelihood of further improvement without surgery.
REFERENCES


